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Decreased bone density in children with Cystic Fibrosis

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Low bone mineral density (BMD), leading to osteoporosis and an increased fracture risk, has been reported in studies using DEXA scan in children with Cystic Fibrosis (CF). Potential factors contributing to low BMD are malnutrition, chronic inflammation and physical inactivity.
Aims: To study BMD (using Quantitative Ultra-sound measurement (QUS)) and the association between BMD and pulmonary function in children with CF.
Methods: Sixty-four children with CF (33 males, mean age (sd) 13.1 yrs (2.9), FEV_1%pred. (sd) 83.3% (23.3)) were measured using QUS (Hologic QDR 4500) on the right calcaneus. Outcome parameters are Broadband Ultrasound Attenuation (BUA, dB/MHz) and Speed of Sound (SOS, m/s), indicators of bone quantity and stiffness. Reference values were obtained from 284 healthy children (118 males, mean age (sd) 12.9 yrs (3.3)). As a pulmonary function test spirometry was used and presented as FEV_1% predicted.
Results: Mean age between CF and the reference group was not significantly different. Body-height (-5.6 cm.; 95% CI: -10.6 to -0.6, $p < 0.01$) and weight (-6.9 kg.; 95% CI: -11.4 to -2.4, $p < 0.01$) were significantly decreased in CF. After adjustment for gender, height and weight, children with CF had a significant decrease in BMD (SOS: -16.1 m/s; 95% CI: -23.9 to -8.3, $p < 0.001$; BUA: -4.1 dB/MHz; 95% CI: -8.1 to 0.0, $p < 0.05$). Within the CF group, a significant positive association was found between FEV_1 and SOS (14.2 m/s; 95% CI: 4.4-24.4, $p < 0.01$), and FEV_1 and BUA (7.1 dB/MHz; 95% CI: 1.7 to 12.5, $p < 0.05$).
Conclusion: Children with CF have decreased BMD (of the calcaneus). Low BMD is associated with poor pulmonary function.

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Upper Extremity Flexibility and Pulmonary Function in Cystic Fibrosis

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The aim of this study was to investigate the relationship between upper extremity flexibility and lung function in patients with cystic fibrosis (CF). Fifteen patients with CF (9-21 years) participated in this study. Pulmonary function test, and inspiratory and expiratory muscle strength (MIP and MEP, respectively) were performed. Functional and lateral reach tests were used to evaluate upper extremity flexibility. Functional reach test score was significantly related with forced expiratory volume in one second (FEV_1, r= 0,54, p= 0,039), forced vital capacity (FVC, r= 0,53, p= 0,045), MIP (r= 0,64, p= 0,011), and MEP (r= 0,59, p= 0,021). Lateral reach test score was significantly correlated with MIP (r= 0,53, p= 0,042). Total flexibility score was significantly related with FVC (r= 0,59, p= 0,019), MIP (r= 0,68, p= 0,006), and MEP (r= 0,52, p= 0,049). In conclusion, upper extremity flexibility was significantly related with effort dependent pulmonary function test parameters and respiratory muscle strength in clinically stable CF patients.

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Pulmonary rehabilitation and quality of life in an adult CF population

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Aims The aim of this study was to assess the effect of a six-week pulmonary rehabilitation (PR) programme on quality of life in an adult CF population in the Mid-Western area of Ireland.
Quality of life is a gauge of the effects of the disease on the various levels of our psychological, psychosocial and physical functioning (Pfeffer et al 2003). Much literature supports exercise programmes to enhance fitness, increase sputum clearance, reduce breathlessness and increase survival in CF (Dodd and Webb1999).
Method Eleven patients undertook the PR programme; four patients completed it. Assessment involved fitness testing using the Modified Shuttle walk test (MSWT), measurement of FEV_1 and quality of life testing via the CF Quality of Life Questionnaire (Gee et al 2000). From the MWST results obtained, a training programme at 70-85% HR max was developed and carried out three times per week for 20-30 minutes (circuits and treadmill). After six weeks of training, patients were reassessed.
Results

	FEV_1 (%)		Litres		Shuttles (#)		VO_2max (ml.min.kg)		Quality of Life (%)	
	PRE	POST	PRE	POST	PRE	POST	PRE	POST	PRE	POST
1	68.7	76	1.941	2.461	58	66	18.69	20.69	75	77
2	91.4	71.4	2.811	2.191	96	102	28.19	29.69	87	94
3	30	44	1.141	1.671	72	110	22.19	31.69	74	86
4	42	49	1.171	1.361	46	73	15.69	22.44	87	94

Please note that patient #2 was suffering from a head cold, leading to a reduction in FEV_1.

Conclusion This study identifies a correlation between FEV_1, fitness levels and QoL but due to the complexity of the disease, PR is difficult to undertake in this patient group. However, Webb and Dodd (1999) reported that patients prefer exercise to any other form of treatment modality, highlighting the role for PR in the physiotherapy management of CF.

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Evaluation of a CF physiotherapy homecare service for adults

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Introduction The Home Physiotherapy Service (HPS) was started in 2004 following a bequest by a patient. It is designed to compliment the homecare service provided by nurse specialists for adult patients living within the Greater London area (300 patients) and is the first Adult CF Homecare Physiotherapy Service in the UK. The service is currently part-time (two days/week), run by two specialist CF physiotherapists. **Aim** To assess patient satisfaction for the service to date. To audit the scope of the current service. **Method** Patient satisfaction questionnaires and information from a computer database were analysed. **Results** 41 patients received 73 visits (range 1-8 visits each, mean 1.8 visits). Average visit time: 51.6 minutes (range 30-150 minutes). To date 59 patients have been referred, the majority by physiotherapists (59%) and homecare nurse specialists (34%). Questionnaire results: 100% preferred home visits and found them useful, 81% felt they had more confidence in carrying out treatment at home, and 72% felt they could discuss more private issues at home. Services provided: clinical assessment, optimization of airway clearance techniques, advice with non-invasive ventilation, exercise programmes, advice with posture, continence issues and inhalation therapy. **Conclusion** The HPS is popular, providing advice and support to patients and carers. There are many paediatric homecare physiotherapy services but as yet this has not been available to the adult population. With the increasing complexity of treatment regimens and longevity of life, physiotherapists are taking a leading role in optimizing patient care in addition to the more traditional model of homecare nurse specialists. The part-time nature of the service means that assistance with routine airway clearance techniques is not provided, this must surely be the next step.